

Desmoplastic fibroma of mandible in a child: case report

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Abstract The aim of this report is to present a rare case of a child with a desmoplastic fibroma (DF) at the mandibular angle. This lesion commonly shows a radiolucent and radiopaque radiographic picture. Due to its aggressiveness and high recurrence rate, an early diagnosis is essential and the complete removal of the lesion is the treatment of choice. We present a case of 11-year-old boy who was referred to be treated for an growth in the left mandibular angle area, a radiolucent and radiopaque feature. After an incisional biopsy, the complete removal of the lesion was done without vascular or nerve damage of the inferior alveolar bundle and without causing mandibular discontinuity. The clinical and radiographic features suggested the diagnosis and allowed the differentiation of DF from other lesions in the maxillofacial area. However, the final diagnosis was only possible through histopathological examination.

Keywords Bone neoplasms · Desmoplastic fibroma · Mandible · Surgical pathology · Jaw neoplasms

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Introduction

Desmoplastic fibroma (DF) is a rare and locally aggressive benign tumor of bone. Histologically, it mimics the desmoid tumor of soft tissues, characterized by collagenization, diffuse hyalinization, cellular monotony with no increase in mitotic activity, low cellularity, stellate fibroblasts and infiltrative margins in most lesions [1]. Radiographically, the lesions are radiolucent, expandable and show a trabeculate appearance, usually associated with a soft tissue mass. DF is more frequently found in long bones and the mandible, followed by the pelvis. However, any bone can be affected [2]. The objective of this article is to report a rare case of DF in the mandibular angle of a child and discuss its most important features.

Case report

A 11-year-old boy was referred to us with a history of progressive asymmetric painless growth at the left mandibular angle. He noticed the alteration six months before the intervention. The patient's general examination revealed no pathological alteration of any other body structure. The patient presented good dental health, with an absence of decay, periodontal disease or oral mucosa alterations.

The panoramic radiograph showed an expandable growth in the left mandibular angle area, without any apparent rupture of cortical bone. The tomograph revealed a radiolucent expandable lesion, with a radiopaque halo adhering to the lingual and buccal cortical and no bone rupture (Fig. 1).

The initial histopathological diagnosis of a DF was obtained by an incisional biopsy. The subsequent surgical removal of the lesion was performed through a submandibular approach, with 702 drill osteotomies, complemented by hammer and chisel. (Fig. 2A) For complete removal of the lesion without vascular or nerve damage, the inferior alveolar bundle was retracted and protected.

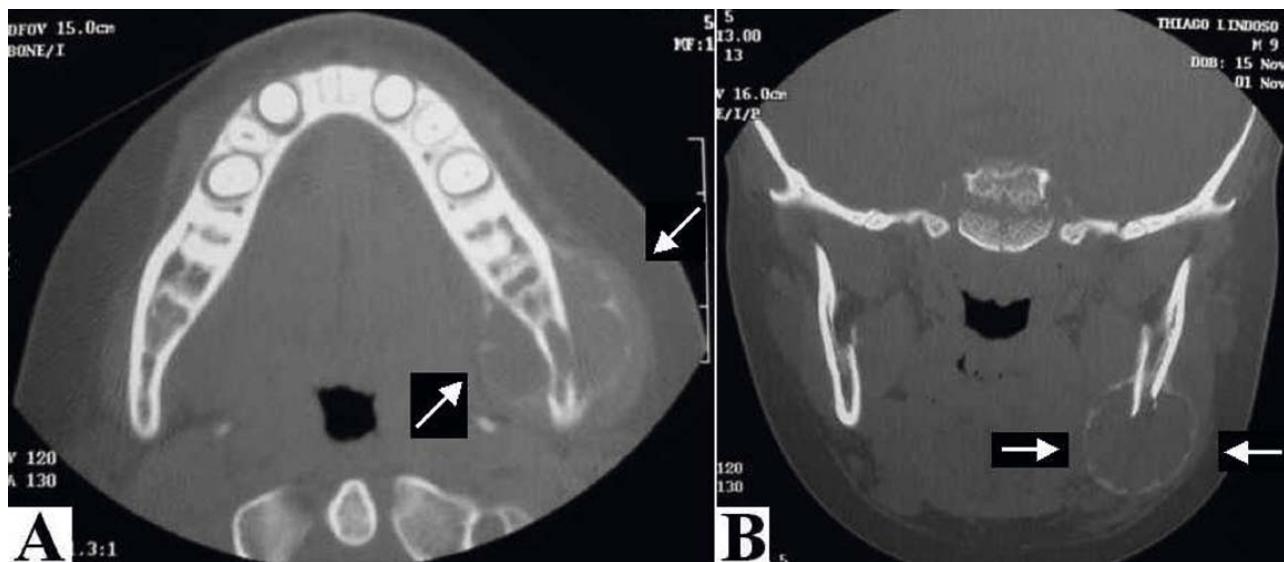


Fig. 1 Preoperative tomographs in axial (A) and coronal cuts (B) where the type of growth and radiographic pattern of the lesion may be seen.

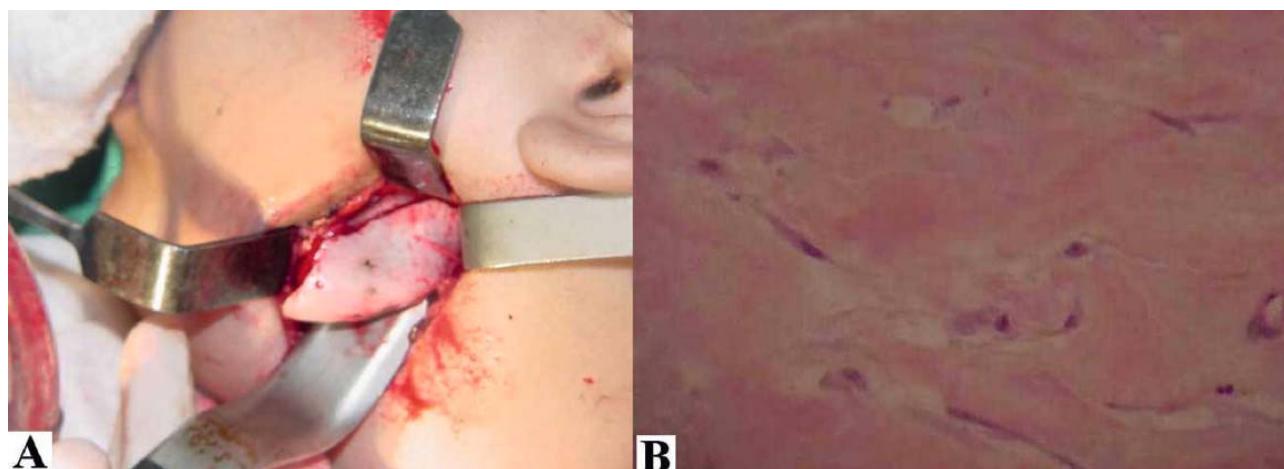


Fig. 2A Transoperative aspect of the removal of lesion. **2B:** Microscopic cuts that evidence the histological pattern of the lesion (hematoxylin-eosin, original magnification x100).

During the postoperative period, the patient showed an injury of the inferior alveolar nerve (neurapraxia). Recovery from this condition occurred after three months. No sign of recurrence of the DF was observed after 33 months.

The final histopathological examination showed, on gross aspect osseous and fibrous components. The soft portion was rubber-like with an irregular form and surface. Microscopically, the histological cuts revealed fragments of dense connective tissue of low cellularity, exhibiting mature spindle-shaped fibroblasts in several fields. The extracellular matrix exhibited intense collagenization and was intensively hyalinized. Thickening of blood vessel walls was present. Fragments of trabeculate bone tissue with normal features completed the histological picture (Fig. 2B).

Discussion

DF is a rare tumor representing 0.06% of all bone tumors and 0.3% of all benign tumors of bone [3]. The tumor is more frequent during the second and third decades of life, usually painless, firm on palpation, and presents a slow growth [4].

Due to the aggressiveness and high recurrence rate of the lesion, the differentiation of DF from other bone lesions is of fundamental importance, because it has a significant impact on the surgical treatment of the lesion [2]. Herford, Reder and Ducic [5] have stated that DF treated with a major excision or resection tends not to reoccur. Tumors treated with simple enucleation or local excisions reoccur

in 20 to 40% of cases, while tumors treated only with curettage reoccur in up to 70% of cases. Intraosseous lesions without signs of cortical rupture or extension to the soft tissues can be treated by curettage. In lesions with signs of aggressive behavior and soft tissue extension, resection has to be considered [4].

Troulis, Williams and Kaban [6] propose that maxillaries tumors in children should be treated by tumor resection, followed by bone graft in a second surgical time and rehabilitation with implants and prostheses.

In our case, the position and the size of the lesion allowed its complete removal without causing mandibular discontinuity. This allowed the preservation of the vascular nervous bundle and no mandibular reconstruction were needed.

In some cases, when the lesion is inoperable or the patient objects to the surgical procedure, the radiotherapy can be used as an alternative. Nag et al [7] interrupt the progression and induced the sclerosis of a DF in the femur of a patient, using 25 radiotherapy sessions during 5 weeks. The future use of radiotherapy in some situations where surgical resection is not possible should clarify the true paper of that kind of therapy in the management of DF.

The radiographic features of DF include a pattern of geographical destruction, trabeculate aspect and bone expansion. Marginal sclerosis and cortical rupture are less common [2]. For Rabin et al [3], the lesion is radiographically characterized by local expansion, well-defined margins and a “soap bubble” appearance. The differential diagnosis of the lesion includes giant cell tumor, aneurysmal and solitary bone cyst, hemangioma, fibrous dysplasia, non-ossifying fibroma and chondromyxoid fibroma [2], odontogenic cysts and tumors, ameloblastoma, myxoma and keratocyst. The radiographic features of the case presented are compatible with the patterns described [2, 3]. Nevertheless, it presents some particularities such as adhesion to the vestibular and lingual cortical with no rupture and the absence of a trabeculate pattern.

Microscopically, DF is characterized by low cellularity, a relatively monotonous appearance of the tumor with an absence of mitotic figures, necrosis and infiltrative margins. The extensive areas of collagenization, diffuse hyalinization and spindle-shape fibroblasts should be emphasized [8]. The histological differential diagnosis of the lesion includes neurofibroma, fibromatosis, sclerotic fibroma of skin, solitary fibrous tumor, myxoma, low-grade fibromyxoid and low-grade myxofibrosarcoma and, in the oral cavity, inflammatory fibrous hyperplasia, fibroma and giant cell fibroma [1, 8]. DF is easily differentiated from malignant neoplasias by the absence of atypical nuclei or mitotic

figures [3]. The immunohistochemical markers are important aids in DF differentiation. DF is immunopositive for vimentin, α -smooth muscle actin (certain cells) and factor XIIIa [8]. The case reported presented all the histological features described.

Every patient with an intraosseous fibrous lesion may be a carrier of tuberous sclerosis. Tuberous sclerosis is an autosomal dominant syndrome that can involve multiple systemic organs. The main manifestations are angiomyolipoma (polycysts of kidneys), heart rhabdomyoma, fibrous dysplasia of bones and facial adenofibroma [1]. The patient described did not present any systemic alterations.

This case illustrates the need to include rare pathological conditions in the initial diagnosis of lesions in the maxillofacial area. It is the clinical and radiographic features that suggest the diagnosis and allows the differentiation of DF from other lesions in the maxillofacial area. Nonetheless, the final diagnosis is only possible by histopathological examination. The treatment indicated in such cases of DF is excision of the lesion.

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